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DOI: 10.4103/pjog.pjog_16_25

Aggressive gliomatosis peritonei associated with mature cystic teratoma: A case report

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Abstract:

Gliomatosis peritonei (GP) is a condition characterized by the dissemination of mature glial tissues throughout the peritoneal cavity. It is usually associated with immature ovarian teratoma but presents with mature cystic teratoma (MCT) in 1% of cases. GP, associated with MCT, is a benign disorder. The majority of cases remain asymptomatic and rarely recur. Here, we present a case of a 22-year-old woman with a history of abdominal enlargement and severe abdominal pain who underwent exploratory laparotomy, peritoneal fluid cytology, bilateral salpingo-oophorectomy, appendectomy, omental biopsy, and Jackson-Pratt drain insertion with histopathologic result of GP with MCT. A month later, the patient had a recurrence of abdominal enlargement, necessitating a second surgery. Immunohistochemistry for histopathologic evaluation and diagnostic imaging are crucial in confirming the diagnosis and guiding the treatment strategy. A multidisciplinary team approach in monitoring and comprehensive support is significant in optimizing outcomes for patients with aggressive GPs associated with MCT. Further research and clinical experience are essential to establish a standardized guideline to improve the management and clinical outcome of this condition.

Keywords:

Gliomatosis peritonei, mature cystic teratoma, reexploration surgery

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Submitted: 29-Apr-2025
Revised: 23-Jun-2025
Accepted: 01-Jul-2025
Published: 29-Sep-2025

Introduction

Gliomatosis peritonei (GP), to date, has been reported to be only about 100 cases worldwide. While commonly linked to immature ovarian teratomas, its association with mature cystic teratoma (MCT) is exceptionally uncommon, estimated to occur in <1% of cases.[1]

The majority of cases of GP were described as having a benign behavior.[2] Glial implants have been considered harmless and may remain stable for long periods, which is associated with a good prognosis. According to a study by Wang *et al*. in 2016, recurrences are frequently encountered in GP-associated immature teratoma and are more susceptible to undergo reexploration surgeries.[1]

We present a case of GP associated with aggressive MCT that necessitated reexploration surgery. This case is significant because it highlights the potential for aggressive behavior in what is typically considered a benign condition.

Case Report

A 22-year-old nulligravid woman presented with a 1-year history of progressive abdominal enlargement. Physical examination revealed a distended abdomen with a palpable abdominopelvic mass.

Contrast-enhanced computed tomography (CT) demonstrated bilateral ovarian masses suggestive of dermoid tumors. Laboratory evaluation showed an elevated serum CA-125 level of 118.5 U/mL, while alpha-fetoprotein (AFP) was within normal limits at 1.5 ng/ml. The patient

How to cite this article: Hamoy LJ, Sia Su ML. Aggressive gliomatosis peritonei associated with mature cystic teratoma: A case report. Philipp J Obstet Gynecol 2025;49:171-6.

was scheduled for bilateral oophorocystectomy with possible salpingo-oophorectomy, depending on intraoperative findings and frozen section results. Intraoperatively, however, the surgical approach was modified and underwent exploratory laparotomy, peritoneal fluid cytology, bilateral salpingo-oophorectomy (BSO), appendectomy, omental biopsy, and placement of a Jackson-Pratt drain. Gross examination revealed that the left ovary was converted to a multicystic mass with serous fluid measuring 20 cm × 18 cm × 15 cm [Figure 1a]. The right ovary measured 12 cm × 10 cm × 9 cm [Figure 1b]. The appendix and omentum seemed congested.

Histopathological examination showed MCT in both ovaries, with the left ovary also exhibiting a mucinous cystadenoma and a focus of small round cell proliferation among mature neural components [Figure 2].

A month after her initial surgery, there was a recurrence of abdominal distention associated with pain, prompting a recommendation for a second surgery. However, the patient was lost to follow-up.

Six months after, she presented with massive ascites associated with severe abdominal pain, prompting a consult. Physical and internal examination noted a 10 cm × 15 cm mobile cystic abdominopelvic mass and a 5 cm × 5 cm cystic mass in the cul-de-sac.

Imaging revealed extensive disease progression. A transvaginal ultrasound revealed multiple loculations within the abdominopelvic cavity, with the largest measuring 12.6 cm × 11.5 cm × 7.8 cm (volume: 591.9 ccs) at the umbilical region [Figure 3] CT scan revealed extensive omental caking, mesenteric thickening, and massive distribution of mucinous material throughout the peritoneal cavity, with multiple mucinous implants causing scalloping of the hepatic and splenic contours suggestive of pseudomyxoma peritonei. Cardiophrenic, lower cervical, and intra-abdominal lymphadenopathies were also noted [Figure 4].

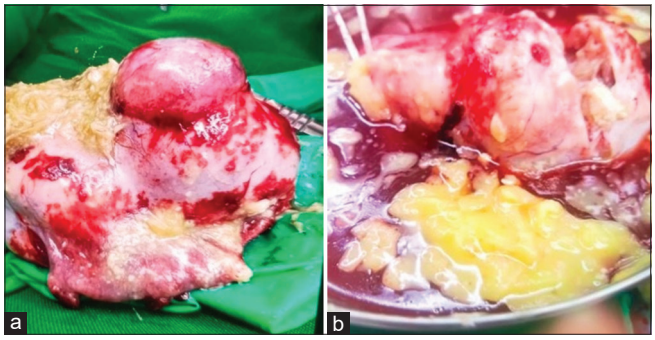

Figure 1: (a) Left ovary was converted to a multicystic with serous fluid 20 × 18 x 15 cm. (b) Right ovary has fat laden contents, hair and sebum, 12 × 10 × 9 cm

Histopathological review of the initial tissues showed MCTs, but the possibility of immature neural tissues could not be ruled out; thus, immunohistochemical studies were done, which confirmed a MCT: Positive for (1) Glial fibrillary acidic protein (GFAP) and (2) S100; Negative for (3) CD68 and (4) leukocyte common antigen [Figure 5].

Repeat tumor markers were normal, with AFP at 2.40 ng/mL, CEA at 0.95 ng/mL, CA 19-9 at 26.8 U/mL, and CA 125 at 25.6 U/mL.

The patient underwent a second operation: preoperative internal jugular vein catheter insertion, cystoscopy, bilateral ureteral stenting, exploratory laparotomy, adhesiolysis, enterolysis, and biopsy of implants.

Intraoperatively, there was 300 cc of serous ascites. Portions of the small intestines and omentum were adherent to the upper anterior abdominal wall, precluding assessment of the liver, gall bladder, sub-diaphragmatic surfaces, stomach, spleen, and kidneys. There were sub-centimeter whitish implants in the accessible portions of the small intestine with a note of retraction along its mesentery. The large intestines were not visualized. The omentum was thickened to approximately 3.0 cm with no gross implants. The abdominal peritoneum was studded with implants, the largest measuring 2.0 cm [Figure 6].

The pelvis had dense adhesions, precluding the assessment of the bladder, uterus, and adnexal area. Residual tumors included the following: Cul-de-Sac

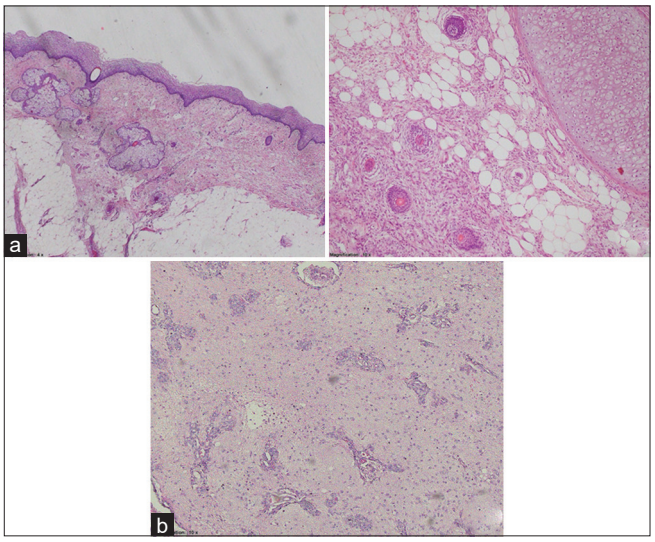

Figure 2: (a) Sections of the ovarian mass showed mature tissue derived from the three germ layers: stratified squamous epithelium with sebaceous glands, glandular epithelium, hyaline cartilage, and hair follicles. (b) There was also small focus of round cell proliferation in a fibrillary background that may was histologically consistent with mature neural epithelium

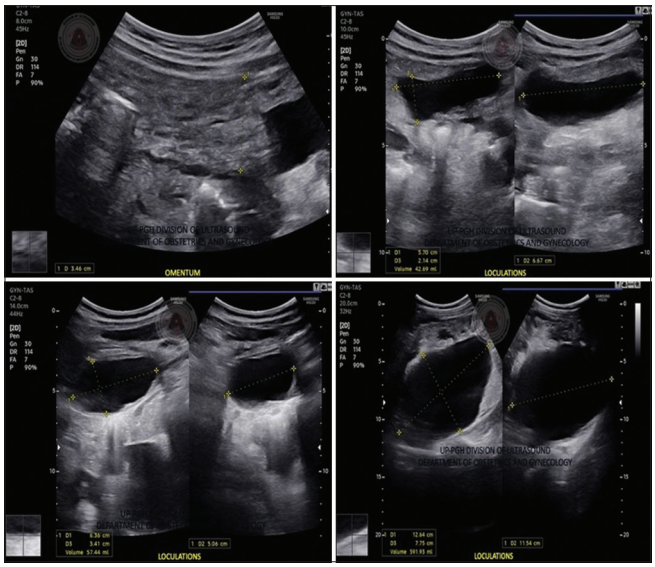

Figure 3: Transvaginal ultrasound. Omentum are variably thickened with non-uniform cystic spaces. Multiple loculations within the abdominopelvic cavity consistent with pseudomyxoma peritonei

mass, small bowel implants, mesenteric implants, and peritoneal implants.

Histologic examination showed sheets of mature glial tissue infiltrating the omental fat, consistent with GP. No areas of immature neuroepithelium or malignant transformation were seen [Figure 7].

Case Discussion

GP is characterized by the implantation of glial tissue on peritoneal surfaces, typically associated with solid ovarian teratomas.[3] It most commonly manifests during the second decade of life and is characterized by miliary-like grayish nodules composed of mature glial and neuronal tissue on the peritoneal surface and omentum. This is generally considered a benign process.

The exact pathophysiology of GP associated with MCT is unclear. There are two theories with regards to the pathogenesis of GP. Some believe that glial implants arose from teratoma through tumor rupture or by metastasis through lymphatics, while others propose that implants arose from pluripotent stem cells. For this patient, it is believed to arise from dislodged or disseminated glial tissue from a surgically ruptured or removed teratoma, leading to glial implants spreading in the peritoneal cavity.[4] GP associated with MCT lesions is extensive, and residual peritoneal disease is common, but patients often remain asymptomatic and inactive for extended periods.

Bentivegna *et al*. documented 10 cases of GP associated with teratoma, with two requiring second surgeries, both associated with immature teratomas as the primary

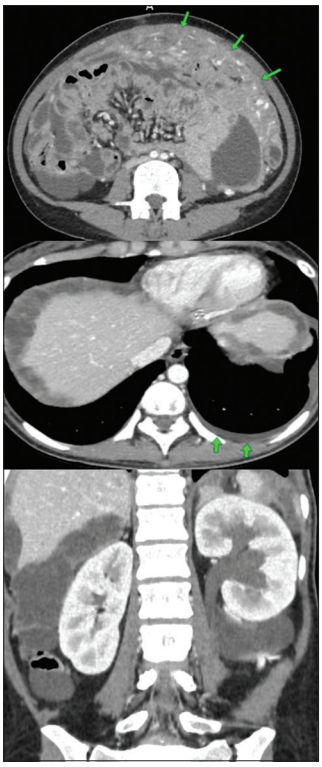

Figure 4: CT Scan of the chest and whole abdomen. Extensive omental caking, mesenteric thickening, and massive distribution of mucinous material throughout the peritoneal cavity. Multiple mucinous implants causing scalloping of the hepatic and splenic contour. Cardiophrenic, lower cervical, and intraabdominal lymphadenopathy. Obstructive uropathy with mild ureteropelvocaliectasia, left

tumor.[5] The study reported a positive outcome for the MCT case, which was asymptomatic, contrasting with our patient, who experienced recurrence only a month after the initial surgery.

While GP is generally considered benign, its association with MCT poses a risk for concurrent malignancy or potential malignant transformation. This was discussed in a study by Bajracharya *et al*., where a patient harbored malignant transformation with squamous cell carcinoma.[4] The study of Wang *et al*. further supported this by focusing on accurate histopathological evaluation to detect malignant elements and the necessity of surveillance of GP associated with MCT patients to prevent malignant change.[1]

GP associated with MCT typically presents asymptomatically but can have an insidious course associated with a pelvic mass, abdominal distention, and pain. This patient experienced early recurrence with an enlarging abdomen and pain.

Imaging plays a major role in the diagnosis of GP, which typically presents as nodular enhancement of the peritoneum.[6] While transvaginal ultrasound may be limited by the size of the nodules, it can serve as an alternative when CT imaging is unavailable. Magnetic

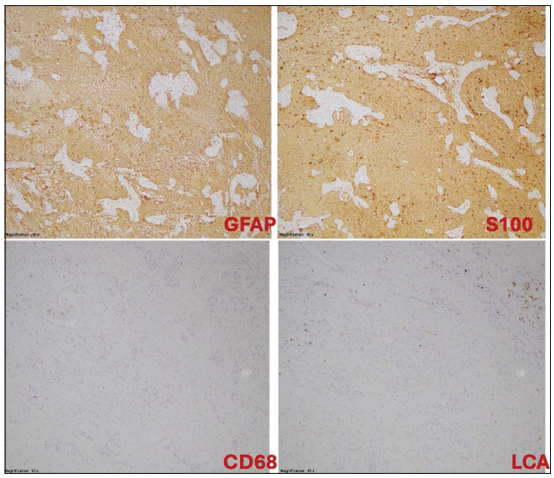

Figure 5: Immunohistochemistry studies showed positive staining for GFAP and S100 protein and negative staining for LCA and CD68, confirming this finding

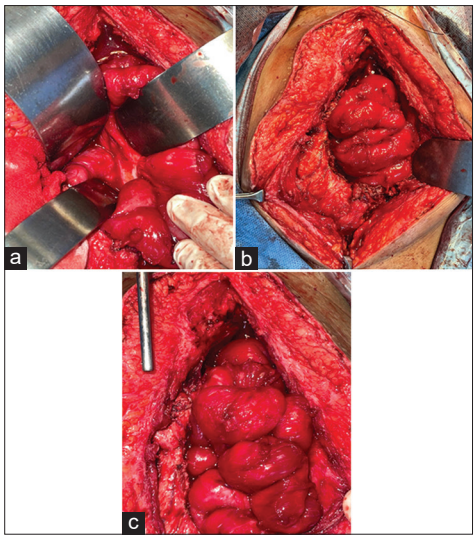

Figure 6: (a) Sub-centimeter whitish implants of the small intestine with note of retraction along its mesentery. (b) The abdominal peritoneum was studded with implants, largest measuring > 2.0 cm. (c) Portions of the small intestines and omentum were adherent to the upper anterior abdominal wall

resonance imaging can also help detect macronodules of glial tissue. In the present case, transvaginal ultrasound showed thickened abdominal peritoneum, matted bowel loops, and massive loculated ascites. CT scan of the abdomen demonstrated extensive ascites with internal septations and a large pelvoabdominal mass characterized by calcified and fat-attenuating components.[6] Based on these findings, peritoneal carcinomatosis was suspected, as it can have clinical and radiological features similar to those of a GP. The transvaginal ultrasound and CT scan showed features suggestive of malignant peritoneal carcinomatosis, although tumor markers (CA-125, AFP, CEA, and CA19-9) were unremarkable.

In a study by Park *et al.*, the researchers explained that imaging features usually have a thickened peritoneum, loculated ascites, and complex adnexal masses, which may imitate malignant peritoneal carcinomatosis.[6] These imaging modalities are crucial for surgical planning and site-specific sampling for confirmatory diagnosis.

The primary management of GP associated with MCT involves surgical intervention, with the goal of maximal resection. The degree of surgical debulking remains debatable, ranging from aggressive approaches to minimize recurrence to more conservative surgeries to reduce operative morbidity. For a mature implant, conservative treatment is indicated; simple surgical excision of the tumor is sufficient, especially given the frequent occurrence of GP during childhood or adolescence. Conversely, when GP with immature teratoma, complete surgical staging and extensive sampling of all implants must be performed. In this patient, initial management included exploratory laparotomy, BSO, appendectomy, and omental biopsy. Despite these measures, fluid reaccumulation and persistent abdominal masses necessitated a second surgery.

Surgical exploration and biopsy were imperative to establish a definitive diagnosis of GP. While GP associated with teratoma is often initially diagnosed on hematoxylin and eosin-stained tissue sections, immunohistochemistry is crucial for differentiating it from low-grade epithelial ovarian tumors. A strong expression of GFAP suggests that tumor cells are mature and well-differentiated.[2] Muller's study provided further insight into the diagnostic criteria for GP. A positive staining reaction of the glial tissue for vimentin and the neural markers GFAP, NSE, and S100, combined with a negative MiB-1, indicated mature nonproliferating tissue, which confirms the diagnosis of a mature glial tissue.[7] The patient's histopathological examination showed MCTs in both ovaries. Immunohistochemical studies were required due to small round cell proliferation in the left ovary, which revealed strong positivity to GFAP and S100. In addition, the omental biopsy revealed mature glial tissue, consistent with GP. This comprehensive histopathological evaluation confirmed the GP associated with MCT diagnosis.

The management of GP associated with MCT presents several challenges, particularly regarding adjuvant therapies. Adjuvant systemic chemotherapy is an accepted treatment in cases of GP associated with immature cystic teratoma. Adjuvant chemotherapy includes bleomycin, etoposide, and cisplatin regimen (BEP) or the vincristine, adriamycin, and cyclophosphamide regimen. However, the limited evidence on the efficacy of chemotherapy for GP associated with MCT complicates treatment decisions.

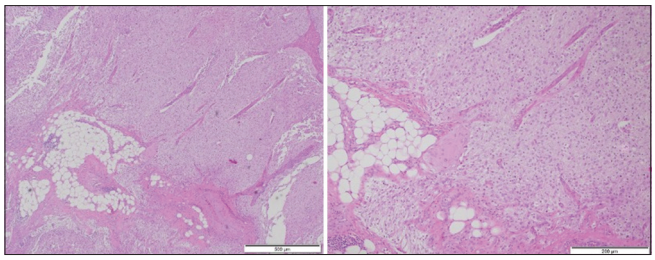

Figure 7: Histologic examination of the omental implant shows sheets of mature glial tissue infiltrating the omental fat, consistent with gliomatosis peritonei. No areas of immature neuroepithelium or malignant transformation were appreciated in the sections examined

Given the benign nature of this condition, traditional chemotherapeutic regimens are not typically indicated, and their role remains controversial.

Recent innovation integrates hyperthermic intraperitoneal chemotherapy with cytoreductive surgery for peritoneal surface malignancies. However, its efficacy in GP cases remains uncertain.

A case report by Liu *et al.* on aggressive GP arising from MCT highlighted the challenges of treatment. In their case, the patient underwent two operations. However, ascites and pain still recurred. Experimental chemotherapy using the BEP regimen was administered due to extensive peritoneal glial nodules, which were difficult to remove completely with surgery. Despite this approach, the patient still had a poor outcome.[2]

In our patient, the histopathological findings showed chronic inflammation and mature neural components without evidence of malignant transformation. These results suggest that nonsurgical therapies may have limited utility. Individualized patient care plans in prioritizing symptom management and improving quality of life are required.

Due to the rarity of GP associated with MCT, there is currently no standard guidance on the duration or methods for patient follow-up. Imaging and tumor marker evaluations may be vital for managing this case.[8]

GP associated with MCT is most commonly observed in adolescent females. Given the typically benign nature and favorable prognosis of this condition, fertility-sparing surgery is often the preferred initial treatment approach.[9] In cases where surgical menopause is induced, hormonal therapy may be considered to manage vasomotor symptoms and mitigate long-term sequelae associated with hypoestrogenism. Estrogen therapy–either alone or in combination with progestin in patients with an intact uterus–is commonly employed. Nevertheless, diligent long-term follow-up remains critical to monitor for recurrence or potential malignant transformation. Comprehensive patient support, including psychological counseling, is vital in addressing the emotional and psychological impact of the diagnosis and the ongoing medical interventions.[10] The psychological burden of watchful waiting, multiple surgeries, and the unpredictability of disease progression is significant. A holistic approach given to this patient is necessary, encompassing not only physical treatment but also psychological and emotional support.

Conclusion

GP associated with MCT remains a diagnostic and therapeutic dilemma, and reporting cases is vital to aid in finding treatment for the recurrence and possible malignant transformation of this condition.

This paper presented a case of an aggressive nature of GP associated with MCT, necessitating multiple surgical interventions. It also highlighted the crucial role of comprehensive diagnostic approaches, including imaging studies and histopathological evaluation with immunohistochemistry, and the need for a multidisciplinary approach in management, monitoring, and psychological support.

This paper presents the difficulties and challenges in managing recurrent GP associated with MCT in the absence of guidelines for this condition.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Authorship contributions

Loryli Jan V. Hamoy, MD – Involved in conceptualization, resources, writing – original draft, review and editing and visualization.

Maria Lilibeth L. Sia Su, MD - Involved in conceptualization, resources, review and editing of the draft and supervision.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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